



Emphysematous pyelonephritis: A case report

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Introduction

Emphysematous pyelonephritis is a rare, necrotizing infection with gas formation of the kidney that primarily affects diabetic patients and carries a high mortality. First described in 1898, there are only approximately 200 cases reported in the literature. Increased awareness and early diagnosis may help improve outcomes.

Case Report

A 65 year-old woman was admitted for new onset diabetes with diabetic ketoacidosis and urinary tract infection. She developed constant, diffuse, abdominal pain, anorexia, tactile fever and chills for 3 days. On admission, temperature was 98.7 F, pulse 113/minute, blood pressure 168/54 mmHg, respirations 49/minute, and oxygen saturation 98% on 5L nasal cannula. She was tachypneic with accessory muscle use, decreased breath sounds and crackles at her left more than right lung base. Cardiac auscultation was normal. Abdomen was distended, diffusely tender, and tympanitic to percussion. She had bilateral costal-vertebral angle tenderness. Initial labs were as follows: Sodium 123 mEq/L, Potassium 4.2 mEq/L, chloride 89 mEq/L, bicarbonate 11 mEq/L, BUN 55 mg/dL, creatinine 1.9 mg/dL, glucose 591 mg/dL, WBC 15.1/cu mm with 57% neutrophils and 32% bands, hemoglobin 14.1 g/dL, and platelets 72,000/cu mm. Urinalysis was notable for specific gravity of 1.020, pH 5.0, glucose > 1000 mg/dL, ketones 15 mg/dL, protein 100 mg%, 2-5 WBC and 5-10 RBC per high power field, and many bacteria. Abdominal radiograph showed dilated loops of bowel but no free air. Chest radiograph revealed an elevated left hemidiaphragm that developed into a left lower lobe consolidation the following day. Renal ultrasound demonstrated a cyst on the right kidney, and the left kidney was poorly visualized. Urine and blood cultures grew out *Escherichia coli*. Abdominal computed tomography scan without contrast was performed revealing gas in and around the left kidney (Figure 1). Platelets dropped to 37,000/cu mm. Left sided nephrectomy was performed and the patient recovered. Pathology revealed severe necrotizing pyelonephritis with hemorrhage and obliteration of the corticomedullary junction and pyramids.

Discussion

Emphysematous pyelonephritis is a necrotizing renal infection characterized by the presence of gas within the renal parenchyma. Reduced blood flow with thrombosis of the kidney may be the initiating event in the pathogenesis of emphysematous pyelonephritis.¹



Figure 1.— Emphysematous pyelonephritis—Class 3B, Arrow depicts extension of gas to pararenal space

Diabetes mellitus is present in 70-96% of patients.^{2,3} Emphysematous pyelonephritis is 3-6 times more common in women.^{1,2} The left kidney (67%) is more frequently involved and bilateral involvement is seen in 12%.^{2,4}

Factors involved in the pathogenesis of emphysematous pyelonephritis include: 1) gas-forming bacteria; 2) high tissue glucose; 3) impaired tissue perfusion; 4) and a deficient immune response.⁵ A vicious cycle ensues with increased gas production causing tissue ischemia, resulting in necrotic tissue, which in turn serves as a substrate for further bacterial proliferation and gas production.^{5,6} Diabetic microangiopathy may contribute to impaired transport of end products and accumulation of gas.⁷ In nondiabetics, lactose may serve as a substrate for fermentation.⁸

Infective organisms can be identified in 92-98% of cases.^{2,9} Organisms frequently associated with emphysematous pyelonephritis are capable of fermenting sugars. Fermentation produces carbon dioxide and hydrogen gases as byproducts.^{5,6,8} Pathogens include *Escherichia coli* (60-90%), *Klebsiella pneumoniae* (26-29%) and multiple others (11%) including *Proteus mirabilis* or *vulgaris*, *Enterobacter aerogenes* and *Pseudomonas aeruginosa*.^{2,3,9} Mixed infections may be responsible for up to 19% of cases.¹⁰ Rare causes include *Streptococcus* species, *Clostridium septicum*, *Candida* and renal amebiasis.^{1,11,12} Other causes of gas within the urinary tract

include recent instrumentation, fistulous connection with a hollow viscous, or penetrating trauma.^{6,13-15}

Clinical Presentation

Typical clinical findings are listed in Table 1.^{2,9,14,16} An abdominal mass may be palpable, although crepitation is rare.¹³ Urinary tract obstruction may be found in 5-40%, while urinary stones are found in up to 27%.⁹ Up to 4% of cases of emphysematous pyelonephritis have polycystic kidney disease.² Other risk factors are drug abuse, neurogenic bladder, alcoholism, and anatomic anomalies.¹⁷

Diagnosis

Due to the lack of specific findings, diagnosis is often delayed. Michaeli et al suggest obtaining a plain radiograph as part of the initial work-up of all poorly controlled diabetics presenting with presumptive urinary tract infection.^{1,14} A plain film detects gas in 33%-85% of cases.^{3,13} Gas seen in the renal parenchyma is diagnostic, but normal appearance does not rule out emphysematous pyelonephritis.¹⁴

Computed Tomography (CT scan) is the diagnostic study of choice.³ It has the highest sensitivity and is useful in demonstrating extent of disease.¹³ Huang et al established a classification system for emphysematous pyelonephritis that is useful in the management and prognosis of this disease (see Table 2).²

Management

Treatment of emphysematous pyelonephritis with antibiotics alone is inadequate. Numerous studies have demonstrated high mortality with conservative management. For class 1 and 2 disease, antibiotics with percutaneous drainage are extremely effective, and can be followed by nephrectomy if the patient does not improve. For classes 3A and 3B, patients are either initially managed with antibiotics and percutaneous drainage or direct nephrectomy depending on the presence of the following four risk factors: 1) thrombocytopenia, 2) acute renal impairment, 3) disturbance of consciousness, and 4) shock. 85% of patients with 1 or fewer risk factors can be successfully treated without nephrectomy. For patients with 2 or more risk factors, only 8% would be cured without nephrectomy, so prompt nephrectomy is warranted.² For patients with class 4 disease patients are usually treated with bilateral percutaneous drainage initially to try to preserve as much renal function as possible.¹⁸ If patients do not improve they must then have nephrectomy.²

Prognostic Features

No significant difference between non-survivor and survivor groups was found in a retrospective study of 38 patients with respect to age, gender, diabetes mellitus history, presence of bacteremia, identity of infecting organisms, blood glucose level, leukocyte count, urinary white blood count, presence or absence of urinary tract obstruction or urolithiasis, and modes of treatment.⁹ Serum creatinine is the most reliable predictor of outcome.⁹ Thrombocytopenia and urinary red blood cell counts are also significant predictors of outcome.⁹ The best predictors of mortality are creatinine > 1.4 mg/dL (sensitivity 93%, specificity 39%), creatinine > 2.5 mg/dL (sensitivity 80%, specificity 70%), platelet < 60,000 x10⁹/L (sensitivity 67%, specificity 87%), platelet < 100,000 x10⁹/L (sensitivity 80%, specificity 61%).⁹

Table 1.— Signs and Symptoms in patients with Emphysematous Pyelonephritis

Finding	Frequency (%)
Fever	56-87
Chills	56-71
Local pain or tenderness	48-71
Shock	29
Lethargy or confusion	19-24
Nausea or Vomiting	16-17
Toxic encephalopathy	16
Dyspnea	13
Abdominal pain w/ dysuria	7
Positive blood culture	42
Positive pus culture	68
Positive urine culture	50

Table 2.— Anatomic Classification System [7]

Class	Description
1	Gas in collecting system
2	Gas in renal parenchyma
3A	Gas or abscess extension to perinephric space
3B	Gas or abscess extension to paranephric space
4	Bilateral, or single kidney

Another more recent study of 48 patients found severe proteinuria (>3g/L), thrombocytopenia (<120 x 10⁹/L), and disturbance of consciousness to be independent factors of poor outcome (poor outcome defined as mortality or need for nephrectomy).²

Overall mortality rate is 18-40%.^{2,3,9} Bilateral emphysematous pyelonephritis results in death in 47-50% of patients.^{1,19} Mortality of patients treated conservatively was 40-80%, 15-60% for those who received percutaneous drainage, 13% for those with initial percutaneous drainage followed by nephrectomy, and 10-30% for those with surgical management.^{2,3,6}

Mortality by classification of Huang et al is as follows: class 1 (0%), class 2 (9%), class 3A (29%), class 3B (19%), and class 4 (50%).² Additionally, failure of percutaneous drainage was as follows: class 1 (0%), class 2 (0%), class 3A (71%), class 3B (30%), and class 4 (75%).²

Conclusion

Clinical trials are lacking due to the low incidence of the disease. Clinicians must have a high index of suspicion for diabetic patients with pyelonephritis. Early imaging should be considered to avoid delays in diagnosis. Optimal management is uncertain but can begin with antibiotics and percutaneous drainage for most cases. Nephrectomy should be performed early for patients with advanced disease, patients with risk factors, or in those who initially failed conservative management options.

See "References" p. 25

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